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Fibrous dysplasia of the spine with sarcomatous transformation: a case report and review of the literature

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Summary. A fibrosarcoma is reported in the spine of a 53-year-old man with polyostotic fibrous dysplasia. There was no history of endocrine disturbances and no previous irradiation. Malignant transformation in fibrous dysplasia is rare. A review of the literature reveals 101 cases of malignant degeneration occurring in fibrous dysplasia. We believe that this is the first report of sarcomatous change arising in an area of fibrous dysplasia in the spine.

Key words: Fibrous dysplasia – Fibrosarcoma – Malignant transformation

Fibrous dysplasia is a common benign pathological condition characterized by fibroosseous metaplasia. There are monostotic and polyostotic forms. Polyostotic fibrous dysplasia may be accompanied by skin pigmentation, endocrine disorders, and precocious puberty, an entity known as Albright's syndrome [1].

Although any bone can be affected, vertebral column involvement is uncommon, expecially in the monostotic type [7, 16, 18, 20]. Thoracic location is even more unusual. Wright and Stoker [20] reported nine cases of vertebral involvement in polyostotic fibrous dysplasia; in only one of these cases was there a lesion in the thoracic region.

Malignant degeneration of fibrous dysplasia is a rare phenomenon. The most common malignant tumor is osteosarcoma (53.4%) followed by fibrosarcoma (17.8%) and chondrosarcoma (8.9%).

Case report

A 53-year-old man complained of back pain and weakness, His blood count, ESR, and alkaline phosphatase level were normal. No previous radiotherapy had been administered. Plain roentgenograms showed expanding lytic lesions involving the thoracolumbar spine and multiple ribs. Computerized axial tomography (CT) and magnetic resonance imaging (MRI) revealed a compressed spinal cord between T7 and T10 (Fig. 1). Biopsy of the left tenth rib revealed typical histopathological changes in fibrous dysplasia. The patient underwent an exploration of his thoracic and lumbar spine through a posterior approach. A decompressive laminectomy between T7 and T11 was done with transpedicular vertebral biopsy, and posterior spinal instrumentation was applied. Histological examination of the specimen from the vertebral body and posterior elements revealed fibrous dysplasia with features of fibrosarcoma (Fig. 2). The patient's symptoms were relieved and his motor function improved. He then received six courses of chemotherapy (Adriamycin). Twenty-eight months after the operation, he was still alive (Fig. 3).

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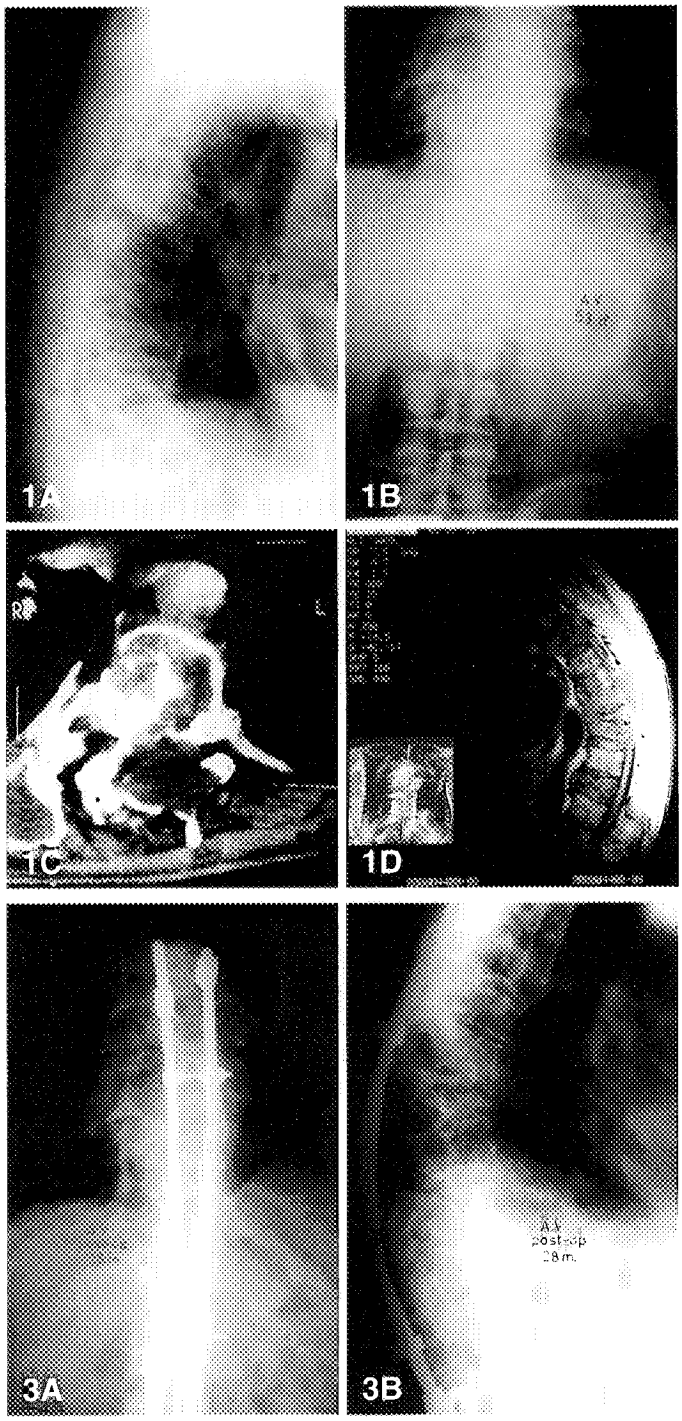
Discussion

Fibrous dysplasia, in either monostotic or polyostotic form, rarely involves the vertebal column. Campanacci [3] reviewed 166 patients and found 2 with monostotic and 2 with polyostotic fibrous dysplasia of the spine. Dahlin and Unni [5] studied 471 patients who had fibrous dysplasia; only 6 patients had vertebral involvement. Schajowicz [14] reported 222 monostotic cases and 36 polyostotic cases, including a sacral lesion in the monostotic group. There is no predilection for any particular portion of the spine. Both the posterior elements and vertebral bodies are affected. When a lesion of the vertebral body becomes extensive, vertebral collapse may occur. Cord compression or nerve encroachment may be caused by expansion of either the vertebral body itself or of posterior elements.

Malignant degeneration of fibrous dysplasia is a rare but well-recognized complication of which 101 cases have been reported [2, 4, 6, 8–13, 15, 17, 19, 21]. The incidence is estimated at 0.4% (6/1517) for fibrous dysplasia and 4% for Albright's syndrome [15]. The common sites were craniofacial bones (35.6%), femur (24.7%), and tibia (12.8%). There has been no other case report regarding sarcomatous transformation in fibrous dysplasia of the spine.

Of the reported cases, 44 patients had monostotic fibrous dysplasia, 46 the polyostotic form, and 11 had Albright's syndrome. There was no predilection for either sex. Osteosarcoma was the most common type of sarcoma that developed in fibrous dysplasia (54 cases). The next most common tumors were fibrosarcoma (18 cases) and



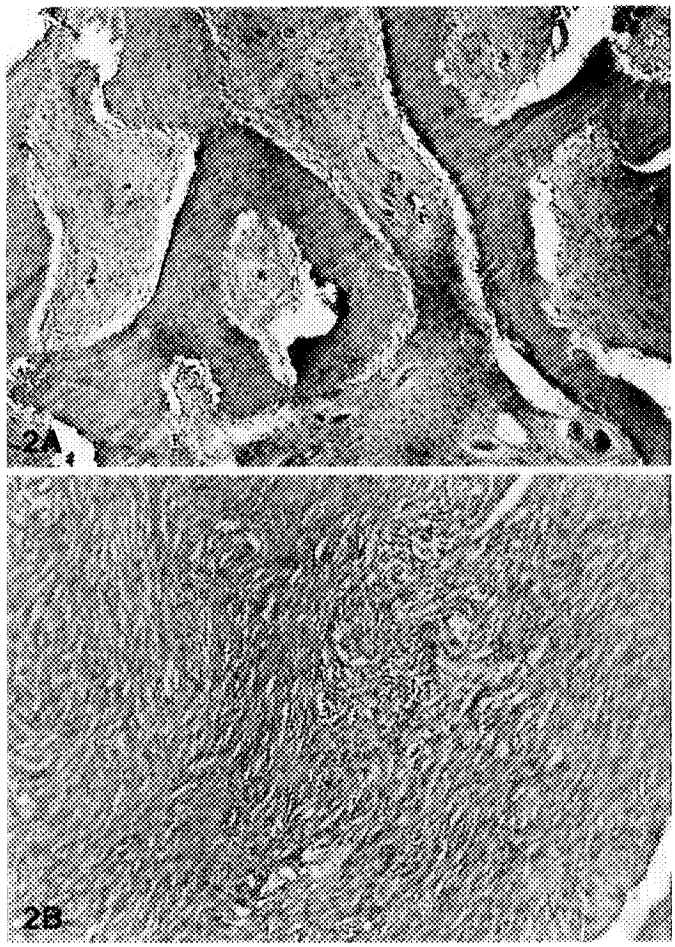


Fig. 1. A, B Anteroposterior and lateral roentgenograms of the thoracolumbar spine show expanding lytic irregular lesions, collapse of the vertebral bodies, and kyphosis. **C** Computerized axial tomogram of the seventh thoracic vertebra. Note the involvement of the body, pedicle, lamina, and the right rib. **D** Magnetic resonance imaging demonstrates compressed spinal cord between T7 and T10

Fig. 2. A Typical histological appearance of fibrous dysplasia. Irregular claw-shaped trabeculae of bone are separated by fibrous connective tissue. Hematoxylin-eosin stain. × 80. **B** Histological appearance of fibrosarcoma. Hematoxylin-eosin stain, × 150

Fig. 3 A, B. Anteroposterior and lateral roentgenograms taken 28 months after surgery

chondrosarcoma (9 cases). Ebata et al. [6] reported a case of polyostotic fibrous dysplasia in which to types of malignant tumor arose: a chondrosarcoma and an osteosarcoma. Malignant degeneration usually develops in the third or fourth decade of life. The average age at onset is 32 years, and the average lag between the development of fibrous dysplasia and sarcoma is 13.5 years [15].

The role of irradiation in malignant degeneration of fibrous dysplasia has been discussed. In the review by Yabut et al. [21], 23 of 83 patients had undergone prior radiation therapy. Chetty et al. [4] reported three cases of malignant neoplasms occurring in fibrous dysplasia involving facial bones. Two of the tumors had been irradiated. The mean interval between radiation therapy and the onset of the sarcoma was 11 years [15]. In this study, 28 patients (27.7%) had a history of irradiation as treatment for fibrous dysplasia. In 20 of them (71.4%), the tumor was an osteosarcoma. It seems that irradiation provokes the fibrous dysplasia to undergo sarcomatous change. Therefore, radiotherapy should not be used for the treatment of fibrous dysplasia.

The case described here is of fibrosarcoma arising in fibrous dysplasia of the vertebral column without prior irradiation. The location of the disease is very exceptional.

As the patient was considered unfit for radical resection of the tumor, he was treated only with posterior decompression and chemotherapy, with good palliative results.

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